COR TRILOCULARE BIAURICULARE

AN UNUSUAL ADULT HEART

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Minor degrees of congenital malformation of the heart are fairly commonly discovered at routine post-mortem examinations and are compatible with normal life and age. Major degrees of congenital malformations are rarer and usually incompatible with long life. However, some who have lived to a good age are recorded.

The earliest imperfect descriptions of bilocular hearts are by Pozzi in 1673 and Lanzoni in 1676. The first fully described case is by Wilson in 1798; he lived for seven days. Many cases have since been described, and Peacock (1866) collected ten. Six cases have been described during this century alone, while Maude Abbott (1927) in her series of 850 autopsies collected seven. The highest recorded age is 16 (Rudolf, 1899).

Trilocular hearts are commoner, the bi-auricular being much more common and more favourable to life. The earliest recorded case is by Chemineau (1699). In Abbott's series fifteen cases are recorded, and there are many more individual reports. Peacock (1866) records the oldest age as 24. In Abbott's series, 35 years for bi-auricular and 31 years for bi-ventricular are given as the maximum ages, but Young (1907) records one, aged 39, and Hedinger (1915) one, aged 56 years.

A common arterial trunk or persistent truncus arteriosus in conjunction with the above abnormalities is even rarer. Only 23 cases have been recorded (Abbott, 1927), and the maximum age recorded by Abbott is 12 years. Vierordt (1898) records one at 16 and another at 19 years.

The following is an extract from the General Register of Deaths in England and Wales, where congenital heart disease was certified to be the *primary cause*.

Age group	1940	1941	1942	Age group	1940	1941	1942
35	17	19	12	60	3	3	8
40	11	11	10	65	1	1	2
45	5	9	8	70	1	1	1
50	6	8	8	75	—	1	—
55	2	2	2				

Unfortunately it does not record the type or degree of malformation.

DESCRIPTION OF CASE

Below is a record of a very unusual case of congenital malformation of the heart—unusual in the age and the gross deformity. Although anatomically a bi-auricular trilocular heart, the auricular septum was membranous and incomplete and therefore functioned imperfectly. There was a single artery from the ventricle (common arterial trunk or persistent truncus arteriosus) and the pulmonary artery probably came as a branch from the innominate artery. There had also been two attacks of theumatic fever with no involvement of the heart.

Mrs. K., aged 56, was admitted to hospital on 10/1/44 with the diagnosis of chronic bronchitis and congenital heart disease with failure. She died the following day.

She had always been blue in the face, and her feet always tended to feel cold. As a child she had

been subject to epileptic fits, " going into jerks and losing consciousness," though never having bitten her tongue nor been incontinent. She had been free of these since her marriage.

How active a life she led is not known. It is known she was married in 1918 and was a widow in 1935. She had no children. In 1944 she was not living with any relatives.

She had previously been in hospital in 1918 with a diagnosis of influenza and mitral stenosis, in 1935 and in 1941 with a diagnosis of congenital heart disease and doubtful acute rheumatism on both occasions.

On examination she was breathless at rest. Cyanosis of hands, lips, and face was present. There was marked clubbing of all her fingers.

The cardiac impulse was diffuse with the maximal impulse in the sixth space in the mid-axillary line. The rhythm was regular. The heart sounds were variable. A blowing systolic murmur conducted into the axilla was heard on admission. No murmurs were heard the next day. In the lungs moist rales were heard all over the chest on both sides. There was an unexplained patch of bronchial breathing over the front right second intercostal space, and behind, between the scapulæ. The urine had a specific gravity of 1012 and a trace of albumen.

The physical signs on her previous admission are of interest. The rhythm was always regular. In 1918 a presystolic murmur at the mitral area was recorded; in 1935 no murmurs were recorded; in 1941 the second sound was accentuated at the aortic area, and a systolic murmur was heard at the third left space and later in the fourth left space. The blood pressure was 136/74 in 1941. Cavernous breathing and bronchial breath sounds were heard at the right apex at the front and back of chest.

The blood count was: hæmoglobin 122 per cent, red blood cells 7,500,000, colour index 0·81, and white blood cells 8400. Unfortunately there is no record of any X-ray or cardiographic examination.

Post-Mortem Examination

The body was that of a small-built woman. Height and weight were not recorded. Except the heart all the organs were on the small side. Signs of congestive failure, with marked congestion of lungs, liver, and kidneys, were present. There was some evidence of chronic bronchitis in the lungs, but it was not very much. There was intense atheroma of the aorta. The left kidney was normal. The right kidney was very small but normal macroscopically and without evidence of arteriosclerotic changes: this was the only other developmental abnormality.

The pericardium was normal and contained a little free fluid. The shape of the heart was triangular with the base superior and the apex inferior. The anterior surface was formed by the ventricle, while the auricle was situated above and behind the ventricle, with two auricular appendices lying one above the other, winding round in a clock-wise direction to the front and ending at the root of the aorta (Fig. 1 and 2). There was moderate hypertrophy of the musculature of both the ventricle and auricles. The heart was not weighed in the fresh state but the weight after fixation in formalin was 310 grammes.

The auricle was incompletely divided by a fibrous septum which ended in a crescentic margin, possibly representing the inferior border of the foramen ovale. Of the two parts, that representing the right auricle was about twice as large as that representing the left. A normal tricuspid valve opened from the larger side and a normal mitral valve from the smaller side (Fig. 3). The right auricular appendix and the wall of the right auricle showed more hypertrophy than that of the left. A superior and inferior vena cava opened into the right auricle, and only two pulmonary veins opened into the left auricle.

The single ventricle represented for the most part that of the left, the right ventricle being represented by a small diverticulum on the right side into which the tricuspid valve opened. There was no sign of any interventricular septum. A single large vessel representing the aorta and guarded by a normal three-cusped valve left the ventricle. There was no pulmonary artery. The coronary arteries took origin from the anterior and left posterior sinuses of Valsalva, the orifice of the right being very large; both arteries showed marked arteriosclerosis and took a somewhat tortuous course on the ventricular surface.

The single vessel arising from the ventricle had no branches until it gave off the innominate artery. The right common carotid artery arose at the junction of the aorta and innominate

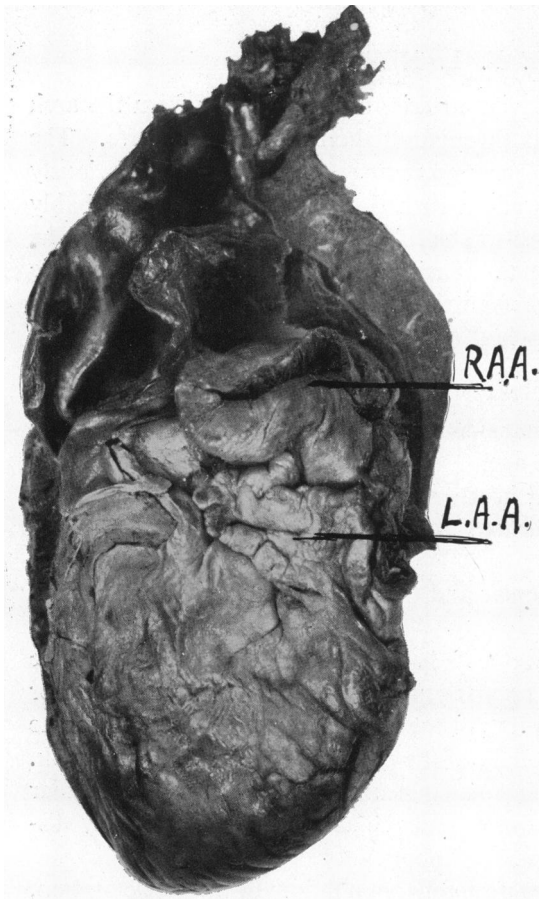

FIG. 1.—Photograph of the heart. R.A.A., right auricular appendage; L.A.A., left auricular appendage.

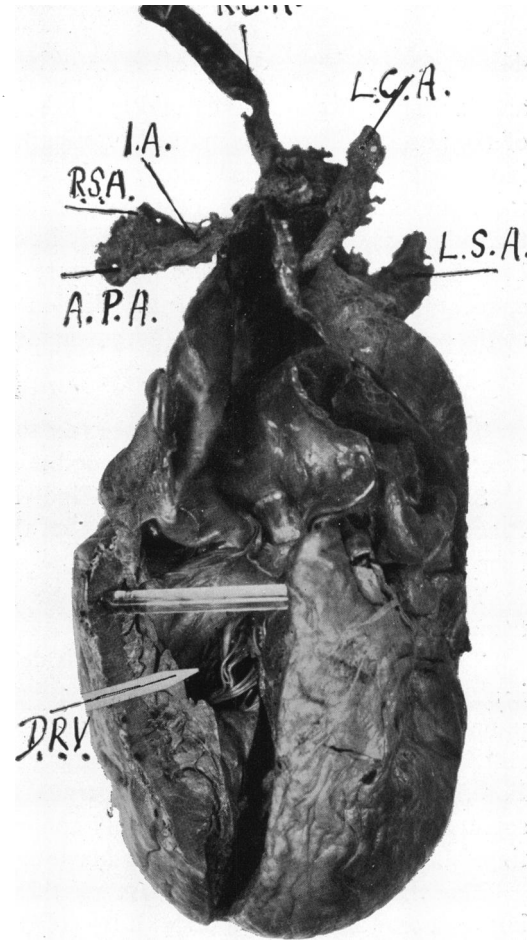

FIG. 2.—The heart opened. R.C.A. and L.C.A., right and left carotid arteries; L.S.A., left subclavian artery; D.R.V., diverticulum representing the right ventricle; A.P.A., aberrant pulmonary artery; R.S.A., right subclavian artery; I.A., innominate artery.

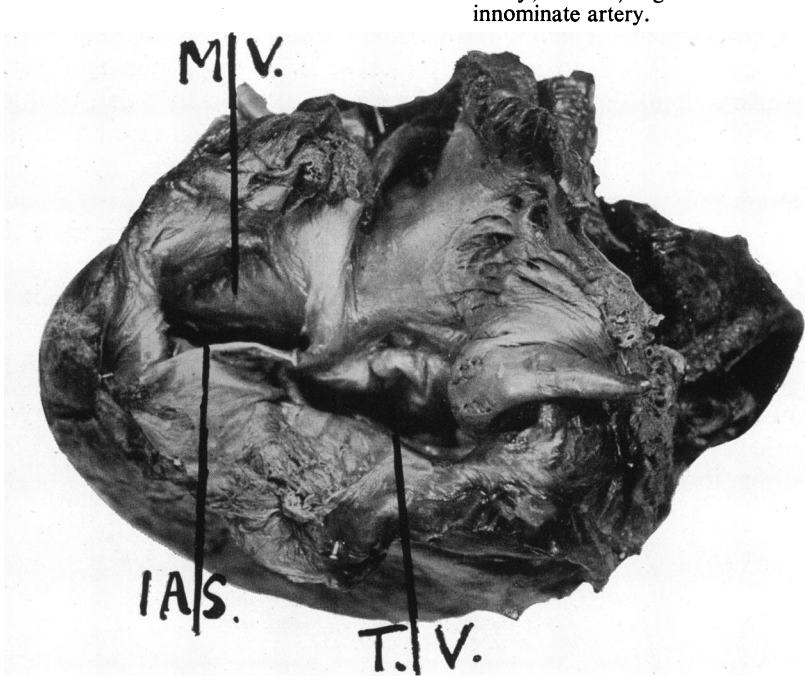

FIG. 3.—Photograph showing the auriculo-ventricular valves. M.V., mitral valve; T.V., tricuspid valve; I.A.S., edge of the incomplete auricular septum.

artery. The left common carotid and left subclavian arteries arose normally from the arch of the aorta. There was marked arteriosclerosis throughout the length of the aorta. The innominate artery divided into two branches, one proceeding outwards being obviously the right subclavian. The other branch appeared to turn downwards and was probably the pulmonary artery, but unfortunately its course was not followed to its termination. The branches from the descending aorta were normal. There was no evidence of rheumatic endocarditis on any of the valves. Section of the ventricular wall showed a patchy fibrosis as the result of arteriosclerosis, but no sign of Aschoff nodes was seen.

Comments

The interest of the case lies in the fact that gross deformity of the heart can occasionally be compatible with normal life to a good middle age.

Such accounts of malformation of individual hearts as are described by Abbott and by Peacock do not show all the features that this heart shows, although each single defect has been described in some heart or other.

This case is the highest age recorded for bilocular and trilocular hearts and for persistent truncus arteriosus.

" When the interventricular septum is defective at its base, as it is in all these cases, there is a tendency for the aortic septum to develop irregularly, thus cutting off a narrow aorta or pulmonary artery, as the case may be, and the calibre of the smaller vessel is likely to become still further reduced in size by the passage of the bulk of the circulation into the larger trunk. For this reason obliteration of one trunk in biloculated hearts, where no interventricular septum is present, is a comparatively common event, and the cases should be sharply distinguished from a true defect of the aortic septum." (Abbott.)

That the single artery in this case was therefore probably not a failure of the aortic septum, but the aorta after separation with loss of the pulmonary artery, is further supported by the fact that there were only three cusps forming the valve. Three or four valves may be present in septal defects of the truncus. The artery arose from the right half of the ventricle. Is it a pulmonary artery turned aorta? A transposition of the vessels or a shift to the right of the vessels is a fairly common finding. Hence probably a true aorta. What about the pulmonary circulation? Unfortunately the pulmonary circulation was never clearly traced post-mortem, but a branch from the innominate artery is presumed to be the pulmonary artery. Various aberrant pulmonary circulations have been described. It is possible that additional blood was supplied to the lungs by the bronchial vessels, but no branch larger than normal was found on the descending aorta. The auricles probably were functionally one although an incomplete septum was present.

A physical sign of note was the area of bronchial breathing over the right chest in the suprascapular region. This observation was made by two independent persons at two different times unknown to each other. No explanation is offered as no post-mortem evidence of any localized lung lesion was found.

Summary

An account has been given of the heart from a woman who lived to be 56 years of age. It was a bi-auricular trilocular heart, but with an imperfect auricular septum. There was a single aorta arising from the ventricle.

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